Disorders of Intestinal Rotation and Fixation

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17.1 Embryology

17.1.1 Normal Intestinal Rotation

In the fourth week of fetal life, the primitive intestine is a relatively straight tube with a slight anterior bulge in the central portion. The superior mesenteric artery arising from the posterior wall enters the center of this anterior bulge. During the next 6 weeks, the intestine grows faster than the coelomic cavity, and as a result it is forced to herniate within the umbilical cord (Fig. 17.1a). The intestine enters the umbilical cord at a point corresponding to the duodenojejunal junction and leaves it at a point corresponding to the primary colonic flexure. The cranial part of the herniation, lying cranial to the superior mesenteric artery, corresponds to the jejunum and ileum as distal as the omphalomesenteric duct, while the caudal part to the terminal ileum and colon. After the tenth week, the coelomic cavity has grown sufficiently and the intestine returns in the abdomen.

Normal intestinal rotation includes a 270° anticlockwise rotation of both the duodenojejunal loop and the cecocolic loop around the axis of the superior mesenteric artery. As a result, the duodenojejunal loop passes from a position above the superior mesenteric artery to a position below and finally to the left side of the artery, while the cecocolic loop passes from a position below the superior mesenteric artery to a position above and finally to the right of the artery (Fig. 17.1a–c).

The process of intestinal rotation has been traditionally divided in three stages. Stage I includes the initial 90° anticlockwise rotation, which will result in positioning of the duodenojejunal loop to the right of the superior mesenteric artery and the positioning of the cecocolic loop to the left of the artery (Fig. 17.1b). This stage occurs during the extracelomic phase of intestinal development. Stage II corresponds to the next 180° anticlockwise rotation, which takes place while the intestine returns into the abdomen and is completed by the end of the 12th week. These further 180° will complete the total 270° rotation, which will finally position the duodenum below the superior mesenteric artery and the duodenojejunal junction to the left of the superior mesenteric artery, as well as the colon above the superior mesenteric artery and the cecocolic loop to the right of the artery (Fig. 17.1c). Stage III constitutes the final 90° anticlockwise rotation of the cecocolic loop, which is usually completed until birth and results in the descent of the cecum from the right hypochondrium below the liver to its final position in the right iliac fossa. Stage III is attributed to dif-

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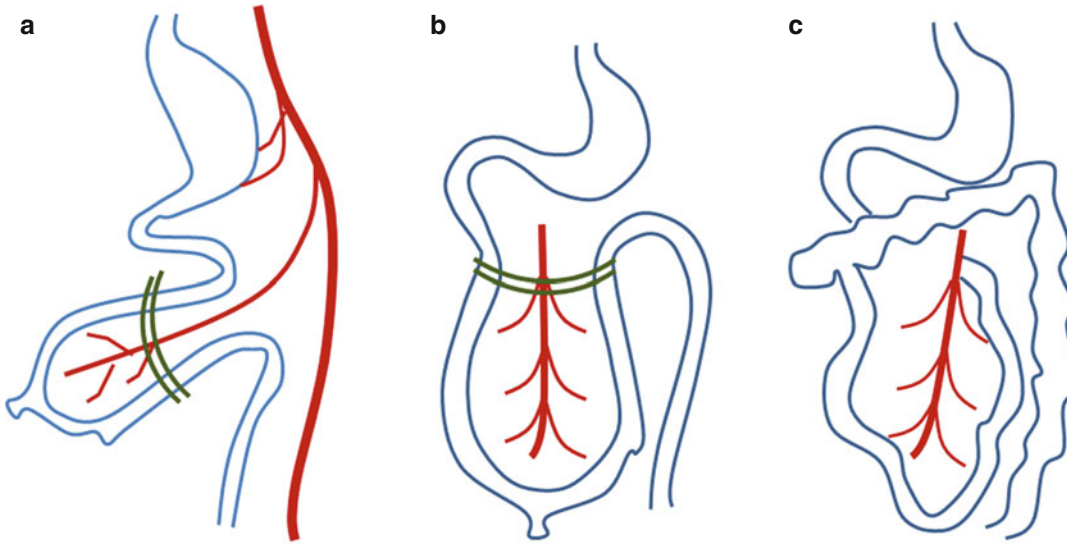

Fig. 17.1 Normal rotation of fetal intestine. (**a**) Orientation of the bowel in the umbilical cord before rotation starts. (**b**) Initial 90° anticlockwise rotation (stage I). (**c**) Further 180° anticlockwise rotation (stage II), which brings the duodenojejunal loop below and to the left of the superior mesenteric artery and cecocolic loop above and to the right of the superior mesenteric artery (From Lister J (1990) Malrotation and volvulus of the intestine. In: Lister J, Irving IM (eds) Neonatal surgery, 3rd edn. Butterworth & Co, London)

ferential growth. It must be highlighted that the staging of intestinal rotation serves only for understanding purposes, while the intestinal rotation is, in reality, a continuous process and should be conceived as such.

After normal rotation and fixation will be completed, the normal mesenteric attachment will extend from the ligament of Treitz, at the level of the pylorus, to the cecum. The second and third portion of the duodenum, as well as the ascending and the descending colon, will be fixed retroperitoneally [1–5].

17.1.2 Disorders of Intestinal Rotation and Fixation

17.1.2.1 Complete Non-rotation

The term refers to the absence of any intestinal rotation. It is characterized by a small and large bowel coursing vertically and a common longitudinal mesentery.

17.1.2.2 Incomplete Rotation

The term refers to the absence of stage II and III rotation, i.e., after the initial 90° anticlockwise rotation of the duodenojejunal and cecocolic loop around the superior mesenteric artery axis, no further rotation has occurred. As a result, the duodenum and small bowel are located on the right side of the artery and the cecum and colon on the left. Congenital adhesive bands between the bowel loops and the parietal peritoneum are often encountered.

17.1.2.3 Malrotation

The term refers to abnormalities occurring during stage II rotation and might include several different types based on the degree of rotation accomplished. In the commonest type, the intestinal rotation has stopped at some point just before the 180°, and, thus, the duodenojejunal loop has failed to cross the midline and lies to the right of the superior mesenteric artery (Fig. 17.2). Similarly, the cecocolic loop has rotated for almost 180° but no further and lies anterior to the duodenum and to the superior mesenteric artery or slightly to the left. Congenital adhesive bands, traditionally known as Ladd's bands, course from the cecum to the parietal peritoneum usually obstructing the second part of the duodenum.

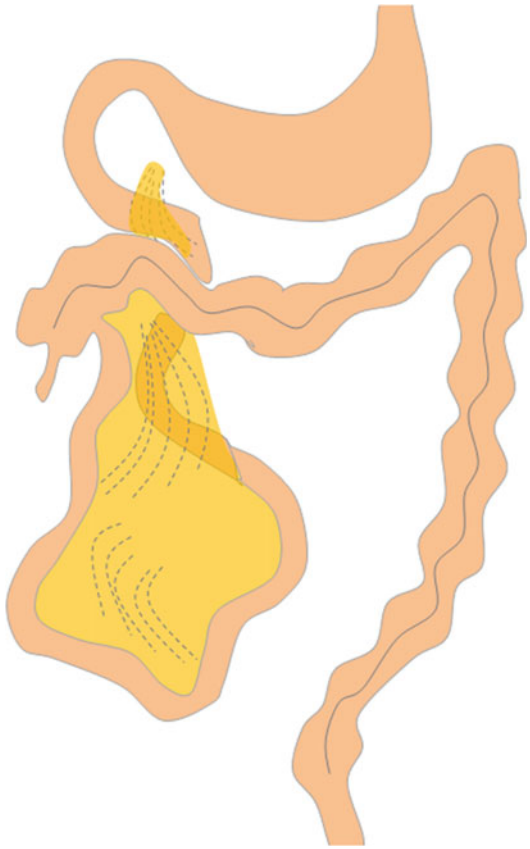

Fig. 17.2 Commonest type of malrotation. Anticlockwise rotation of duodenojejunal and cecocolic loops has stopped at 180° (From Lister J (1990) Malrotation and volvulus of the intestine. In: Lister J, Irving IM (eds) Neonatal surgery, 3rd edn. Butterworth & Co, London)

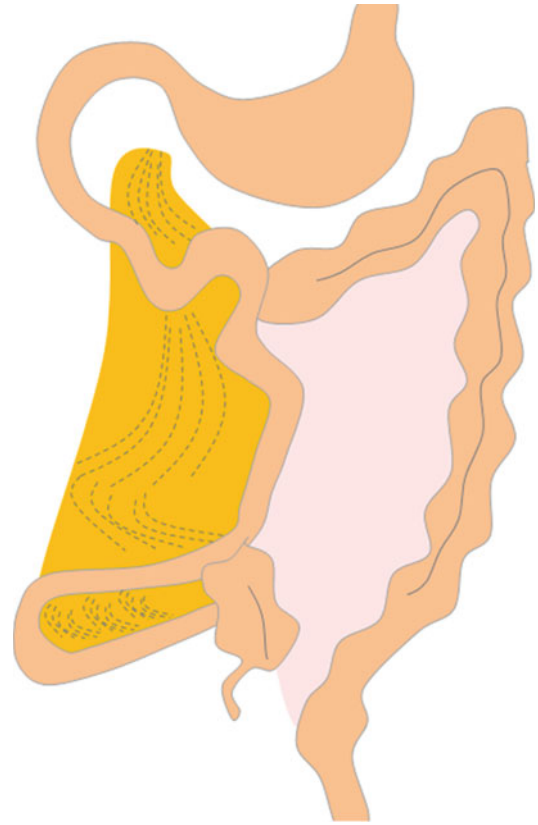

Fig. 17.3 Reverse rotation following the initial anticlockwise 90° rotation of stage I. The 90° clockwise intestinal rotation (From Lister J (1990) Malrotation and volvulus of the intestine. In: Lister J, Irving IM (eds) Neonatal surgery, 3rd edn. Butterworth & Co, London)

17.1.2.4 Reverse Rotation

It refers to less common types of malrotation, in which the first 90° anticlockwise rotation (stage I) is followed by further 90–180° in the clockwise direction. As a result, the duodenum lies anterior to the superior mesenteric artery. The position of the cecum varies, based on the degree of its rotation. It might descend in the lower abdomen behind the small bowel mesentery, after a 90° clockwise rotation (Fig. 17.3), or it might cross the midline behind the superior mesenteric vessels and reach the right iliac fossa—after a 180° clockwise rotation (Fig. 17.4).

17.1.2.5 Atypical Malrotation or Malrotation Variant

The intestinal rotation has been interrupted at some point between 180 and 270°. As a result, the ligament of Treitz is to the left of the midline but lower than the level of the pylorus.

In most cases of abnormal intestinal rotation and fixation, the small intestine has a narrow mesenteric base and, therefore, is prone to twist around the mesenteric vessels on a clockwise direction causing midgut volvulus. Other causes of intestinal obstruction in malrotation are the kinks and compression of the lumen caused by the congenital bands.

The lack of normal fixation of the right or left colon might result in the formation of potential spaces within the mesocolon, like hernial sacs, in which the small intestine might be entrapped causing right or left mesocolic hernias (Fig. 17.5) [6]. These internal hernias might cause recurrent partial obstruction of the small bowel or might eventually result in complete obstruction and strangulation [7–16].

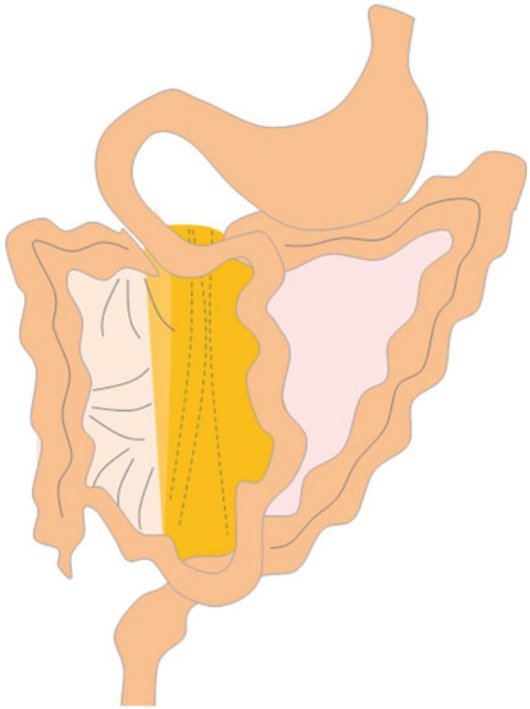

Fig. 17.4 Reverse rotation following the initial anticlockwise 90° rotation of stage I. Complete clockwise intestinal rotation through 180° (From Lister J (1990) Malrotation and volvulus of the intestine. In: Lister J, Irving IM (eds) Neonatal surgery, 3rd edn. Butterworth & Co, London)

17.2 Epidemiology and Associated Malformations

Intestinal malrotation is seen in up to 1:6000 live births. Most patients with volvulus (52–64 %) present in the first month of life and the majority of them (70 %) within the first week of life. Sporadic cases occur throughout life. Associated anomalies are found in 30–60 % of cases of malrotation. These include diaphragmatic hernias, exomphalos and gastroschisis, complete duodenal atresia or duodenal webs, jejunal atresia, Hirschsprung's disease, anorectal malformations, and mesenteric cysts. Rotation and fixation abnormalities are also known to coexist with heterotaxy, with 70 % of these patients having malrotation [9, 10, 17, 18].

17.3 Clinical Manifestations

The primary symptom is the sudden onset of forceful vomiting, usually bilious and less commonly yellowish, in a previously healthy infant. Vomiting might be due to duodenal obstruction caused by Ladd's bands, in the absence of volvulus, or might be the result of an acute obstruction caused by midgut volvulus. Crampy abdominal pain is also common. The obstruction might not be complete, so meconium and stool may be passing. Blood-stained emesis and passage of blood per rectum suggest bowel strangulation and are ominous signs.

Abdominal distention may or may not be present. Sometimes, the upper abdomen might appear distended and the lower abdomen scaphoid. Generalized distention is usually indicative of gangrenous volvulus and is a late sign. Tenderness on palpation is not a constant finding and, like abdominal distention, might be a late sign. Dehydration secondary to vomiting and/or intestinal ischemia, metabolic acidosis, and sepsis develop rapidly.

Intermittent or partial chronic midgut volvulus usually presents in children older than 2 years with symptoms of chronic vomiting, usually bilious, intermittent colicky abdominal pain, hematemesis, diarrhea, constipation, failure to thrive, and protein-calorie malnutrition.

In the rare types of reverse rotation, in which the colon lies behind the superior mesenteric artery, symptoms associated with partial or complete colonic obstruction might present later in adult life [19–21].

17.4 Radiologic Diagnosis

A plain abdominal film showing gaseous distention of the stomach (Fig. 17.6a), and sometimes of the proximal duodenum as well (Fig. 17.6b), and a relatively gasless pattern in the rest of the abdomen is typical of midgut volvulus. Such a film might be all what is required to make the diagnosis in an infant with bilious vomiting, who is not stable enough to undergo an upper GI contrast study. However, plain abdominal radiog-

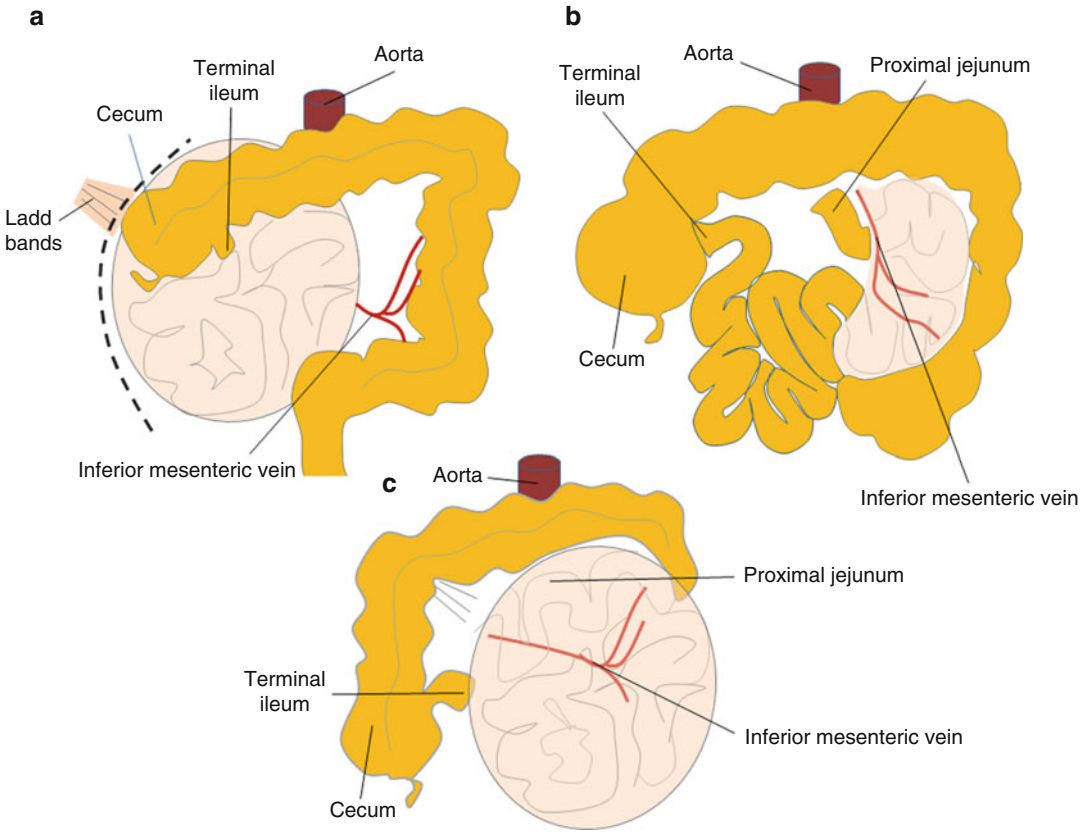

Fig. 17.5 *Right* (**a**) and *left* (**b**, **c**) mesocolic hernias. The interrupted line in (**a**) shows the surgical incision at the area of the lateral peritoneal reflection used to reduce the herniated small bowel from the mesocolon. Note that the inferior mesenteric vein delineates the right margin of the left mesocolic hernias in (**b**) and (**c**) (From Willwerth BM et al. (1974) Congenital mesocolic (paraduodenal) hernia: embryologic basis of repair. Am J Surg 128:358)

raphy may not always be diagnostic, and, thus, an upper GI contrast study should always be considered in a bilious vomiter with a normal abdominal film.

The upper GI contrast study is the investigation of choice for the evaluation of a patient with suspected abnormalities of intestinal rotation. In normal intestinal rotation, the duodenum descends to the right of the midline, courses transversely to the left, and then ascends to the left of the midline up to duodenojejunal junction at the level of the pylorus; the loops of the proximal jejunum are subsequently seen on the left of the midline.

In malrotation, the duodenum descends to the right of the midline and fails to course transversely to the left of the midline (Fig. 17.7a). Instead of crossing the midline, the contrast medium is seen to fill the jejunal loops on the right of the midline (Fig. 17.7b). Delay in the passage of the contrast into the jejunal loops and the characteristic spiral configuration of the proximal jejunum, known as "corkscrew" sign, indicate obstruction because of midgut volvulus (Fig. 17.8). In atypical malrotation, the duodenojejunal junction is demonstrated at the midline or to the left of the midline but lower than the level of the pylorus (Fig. 17.9).

Although contrast enema is considered of low diagnostic value and is in present times rarely used, it has historically been used and can theoretically demonstrate the displaced cecum. However, 20–30 % of the cases with malrotation have a normally sited colon, and, furthermore, the mobile cecum is frequently seen in the right

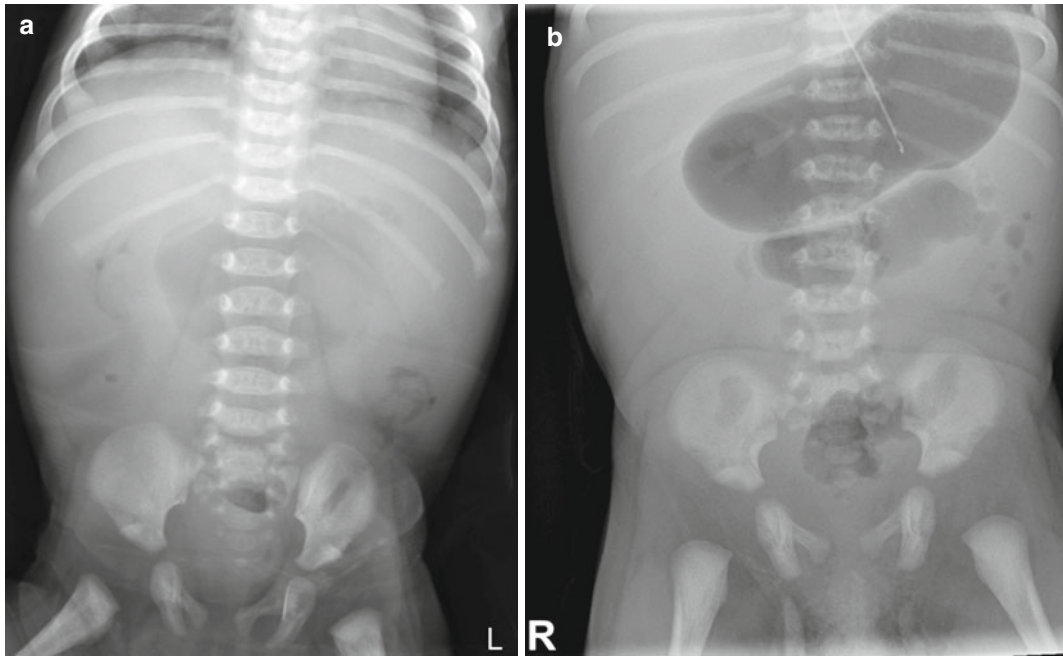

Fig. 17.6 Plain radiographs of infants with midgut volvulus, showing gaseous distention of the (**a**) stomach and of the (**b**) stomach and duodenum, with a paucity of gas distally

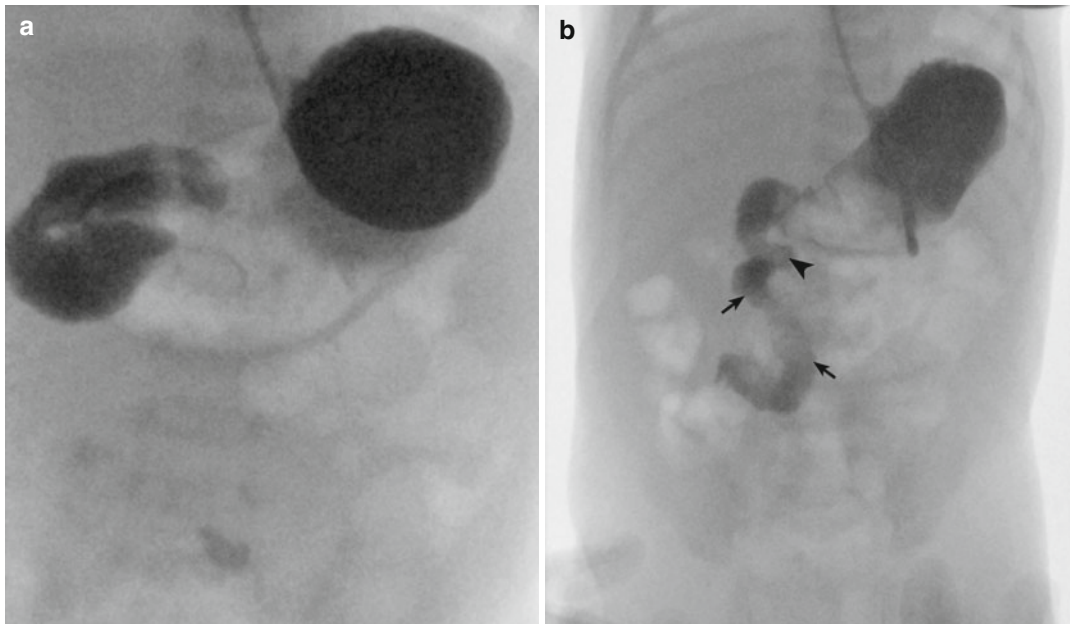

Fig. 17.7 Images from contrast studies of infants showing malrotation. (**a**) The duodenum descends to the right of the midline and then fails to course transversely to the left. (**b**) The duodenojejunal flexure (*arrowhead*) lies to the right of the midline, and the contrast is seen passing from the duodenum to the proximal jejunum (*arrows*) to the right of the midline

Fig. 17.8 Contrast study of an infant with midgut volvulus. The proximal jejunum courses inferiorly, to the right of the midline, in a spiral configuration

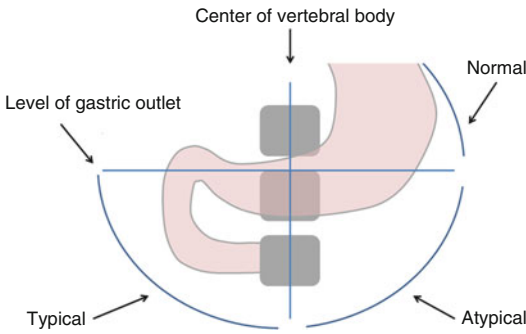

Fig. 17.9 Classification of malrotation as typical and atypical on the basis of the location of the ligament of Treitz (and duodenojejunal flexure) in relation to the midline, as indicated by the vertebral body, and to the level of the gastric outlet (From Dassinger MS, Smith SD (2012) Ch 86. Disorders of intestinal rotation and fixation. In: Coran AG et al. (eds) Pediatric surgery, 7th edn. Elsevier Saunders, Philadelphia)

iliac fossa. The converse is also true, with the position of the cecum in normal individuals being variable [22, 23].

Sonography has lately become popular and is increasingly used to demonstrate abnormalities of intestinal rotation by showing an abnormal relationship of the superior mesenteric vein to the superior mesenteric artery. The inversion in the superior mesenteric artery and vein relationship, with the superior mesenteric artery on the right and superior mesenteric vein on the left, anterior to the abdominal aorta, on color Doppler imaging is diagnostic of malrotation. In some cases of malrotation, however, the orientation between the vessels is normal. Additional sonographic signs are a fluid-filled dilated duodenum and transverse intraperitoneal duodenum. The swirling appearance of the mesentery and superior mesenteric vein around the superior mesenteric artery, known as "whirlpool" sign, on color Doppler imaging is suggestive of midgut volvulus [24, 25].

17.5 Management

Rapid intravenous fluid resuscitation, initiation of parenteral broad-spectrum antibiotics, placement of a nasogastric tube, and expeditious laparotomy are essential.

17.5.1 Ladd's Procedure

The abdomen is opened through a supraumbilical right transverse incision. Ladd's procedure consists of the following steps: (1) exteriorization of the bowel and inspection of the bowel and the mesenteric root in order to recognize the type of the malrotation, (2) counterclockwise derotation of the volvulus, (3) division of the Ladd's bands and straightening of the duodenum, (4) appendicectomy, and (5) placement of the small bowel in the right abdomen and of the cecum in the left lower quadrant.

Exteriorization of the entire bowel is necessary, avoiding traction to the mesentery, in order to understand the anatomy. The volvulus is usually untwisted in 180° turns until the transverse colon and cecum are brought into view anterior to the superior mesenteric pedicle. The bowel is covered with warm moist packs until its pink color returns. Common findings

suggestive of malrotation include Ladd's bands extending from the terminal ileum, cecum, and/or right colon to the duodenum, fixation of the duodenum and/or proximal jejunum to the cecum and/or right colon, and abnormal positioning and mobility of the cecum and/or the duodenum. In the commonest type, the cecum is found in the right hypochondrium, fixed by Ladd's bands passing over the second and third parts of the duodenum (Fig. 17.2). The duodenum should be adequately freed from all adhesive bands until it is straightened and seen to course freely downward to the right side of the abdomen (Fig. 17.10). Dissection is carried out close to the serosa of the duodenum with careful attention to the superior mesenteric vessels. All additional adhesive bands between intestinal loops or between loops and the parietal peritoneum should be meticulously divided until the whole intestine from the duodenum to the sigmoid colon is free of kinks, in order to avoid recurrence of obstructive symptoms. Appendicectomy should not be performed if the cecum appears severely cyanotic.

It is important to prove that no associated intrinsic duodenal obstruction is present. For this reason, passing a nasogastric tube through the duodenum or a feeding tube through a gastrotomy is recommended.

In reverse rotation, it is sometimes possible to move the colon from behind the mesenteric vessels to a position anterior to them by a 360° anticlockwise rotation of the midgut. However, freeing the duodenum and the underlying mesenteric vessels from the colonic wall is usually sufficient to release the colonic obstruction.

In the presence of a mesocolic hernia, the hernial sac should not be resected, because this might cause devascularization of the colon. In the right mesocolic hernia, the entrapped small bowel is freed by incising the avascular lateral peritoneal reflection of the right colon (Fig. 17.5a). In the left mesocolic hernia, particular attention should be drawn to retain the inferior mesenteric vein, which courses along the right margin of the hernial sac (Fig. 17.5b, c). The small intestine is sometimes easily reduced through the neck of the sac without the need for further dissection.

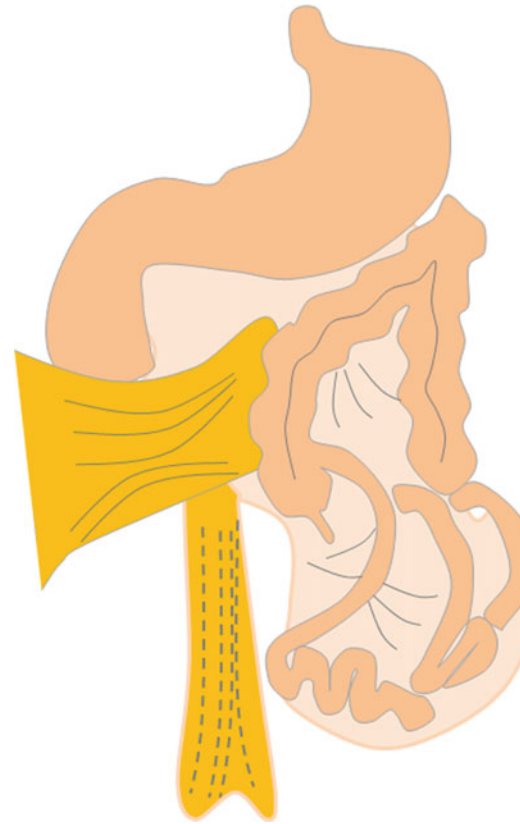

Fig. 17.10 Division of Ladd's bands in order to release and straighten the duodenum (From Lister J (1990) Malrotation and volvulus of the intestine. In: Lister J, Irving IM (eds) Neonatal surgery, 3rd edn. Butterworth & Co, London)

However, it is occasionally required to mobilize the inferior mesenteric vein and/or even make an incision to the right of the vein in order to widen the neck of the sac. After reduction of the mesocolic hernias, the peritoneum of the sac is sutured to the posterior wall to obviate recurrence.

When there is a localized gangrenous segment, then segmental resection and primary anastomosis should be performed. When, however, there are multiple areas of questionable viability or when the entire midgut appears nonviable, the tendency should be on the side of underresection rather than over-resection. A relook laparotomy 12–24 h later, when recovery of the affected bowel loops and/or demarcation of necrotic bowel segments will be clearly evident, is recommended. Preserving the minimum length

of the intestine required for survival, preferably including the ileocecal valve, should be the highest priority.

Operative correction of asymptomatic patients found to have atypical malrotation has been recently questioned. Atypical malrotation is less likely to be complicated with volvulus or internal hernias in comparison to typical malrotation (2 vs. 16 % and 7 vs. 21 %). Moreover, patients with atypical malrotation more commonly present with persistent symptoms post-Ladd's procedure in comparison to patients with typical malrotation (21 vs. 12 %) [26, 27].

17.5.2 Laparoscopic Approach

Minimally invasive techniques can be used for both diagnosis and management of malrotation and might be particularly useful in cases with doubtful diagnosis. The comparative studies between the open and laparoscopic approach are limited by their retrospective nonrandomized design. Theoretically, although laparoscopy has well-recognized benefits, it is also believed to cause fewer adhesions and, as a result, to be possibly related with a higher risk of recurrent volvulus. No significant differences in terms of postoperative complications between the laparoscopic and open approach have been revealed to date. Prospective trials with long-term follow-up are required to make safe conclusions [28–31].

17.5.3 Postoperative Complications

The dilatation of the duodenum and the vascular compromise of the bowel might cause prolonged postoperative ileus even up to 5–7 days. This is managed with expectant policy, continuous gastric aspiration through a nasogastric tube, and intravenous fluids. Patients with extensive bowel injury and short bowel syndrome will need special management with long-term total parenteral nutrition.

The incidence of postoperative intussusception is 3.1 % (vs. 0.05 % in other laparotomies). It usually presents with abdominal distention and bilious vomiting 5–8 days postoperatively [32]. The incidence of postoperative adhesive ileus is 4 %. The incidence of recurrent volvulus is low (0.5–1.3 % in reported series).

Mortality rate has reduced significantly over the last 60 years. However, it is still at least 65 % when more than 75 % of the bowel is necrotic.

17.5.4 Management of Asymptomatic Malrotation

Controversy exists over the management of asymptomatic, typical or atypical, malrotation, which has usually been diagnosed incidentally during the evaluation for nonspecific complaints or prior to reflux surgery. Moreover, atypical malrotation is at a significantly lower risk of volvulus and internal hernia in comparison with typical malrotation and is associated with increased incidence, as high as 13 %, of persistent symptoms postoperatively—when symptomatic—and increased incidence, as high as 22 %, of postoperative complications [33].

17.5.5 Management of Malrotation in Heterotaxy Syndrome

Management of asymptomatic malrotation in heterotaxy syndrome remains also controversial. Because of the high incidence of malrotation in heterotaxy, screening of all these patients and prophylactic Ladd's procedure in the presence of malrotation are the standard of care in many centers. However, high complication rates up to 14 %, including a 10 % incidence of small bowel obstruction, have been reported after Ladd's procedure in heterotaxy patients. On the basis of findings of increased morbidity and mortality after Ladd's procedure in heterotaxy patients, some authors now suggest that routine screening of asymptomatic heterotaxy patients for malrotation should be abandoned. Others have suggested that patients with left atrial isomerism are less likely to be malrotated than patients with right atrial isomerism, and, therefore, they should be offered expectant management if they are asymptomatic [34, 35].

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